Cureus

Open Access Case Report

DOI: 10.7759/cureus.49490

A Rare Case of Benign Long-Standing Ecchordosis Physaliphora

Yahea Alzahrani [1]

1. Department of Internal Medicine, Taif University, Taif, SAU

Corresponding author: Yahea Alzahrani, dr.yahea@gmail.com

Review began 11/18/2023
Review ended 11/22/2023
Published 11/27/2023

© **Copyright** 2023
Alzahrani. This is an open access article distributed under the terms of the Creative Commons Attribution License CC-BY 4.0., which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Abstract

Ecchordosis physaliphora (EP) is a rare benign lesion arising from embryonic notochordal remnants, typically located in the retroclival region. This case report presents a 46-year-old male patient experiencing intermittent headaches and occipital pain. Imaging revealed a well-defined, smoothly corticated bony lesion on the left side of the clivus, accompanied by a characteristic bony stalk devoid of any aggressive features. A review of the patient's medical records indicated stable imaging findings of the lesion over six years. Clinicians and radiologists should be familiar with EP as a benign entity and differentiate it from aggressive pathologies.

Categories: Neurology, Neurosurgery, Radiology
Keywords: ct, mri, chordoma, notochord, ecchordosis physaliphora

Introduction

Ecchordosis physaliphora (EP) is a rare congenital benign hamartomatous lesion originating from nodal cord remnants [1]. It is most commonly found in the retroclival prepontine region of the middle cranial fossa but can also be found anywhere along the midline from the skull base to the sacrum [2]. Ecchordosis physaliphora is typically asymptomatic but can occasionally cause mass effects with compression of the brain stem or cranial nerves [3].

Histopathologically, ecchordosis physaliphora is quite similar to chondroma [3]. The characteristic imaging findings of ecchordosis physaliphora include a well-demarcated, smoothly corticated bony defect in the clivus on computed tomography (CT) and a hypointense lesion on T1-weighted images and a hyperintense lesion on T2-weighted images, with no contrast enhancement on magnetic resonance imaging (MRI) [4].

A case of ecchordosis physaliphora is presented in this article with classic imaging characteristics, including the pathognomonic osseous stalk in both CT and MRI, with no change in size and appearance over a period of six years.

Case Presentation

A 46-year-old male patient presented with intermittent headache and occipital pain. The patient was afebrile with stable vital signs upon initial evaluation. Laboratory tests showed no abnormalities, and the neurological examination was unremarkable. A non-enhanced brain CT (Figure *1*) demonstrated a clearly defined osseous defect on the left side of the clivus. The margin of this defect is well-defined, smooth, and devoid of aggressive characteristics. A tiny, hyperdense stalklike structure, representing a pathognomonic osseous stalk, is also visible projecting from the clivus.

How to cite this article
Alzahrani Y (November 27, 2023) A Rare Case of Benign Long-Standing Ecchordosis Physaliphora. Cureus 15(11): e49490. DOI 10.7759/cureus.49490

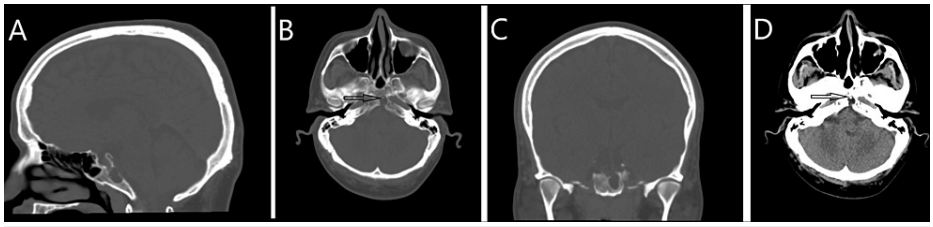


FIGURE 1: Non-enhanced brain CT

Sagittal (A), axial (B), and coronal (C) CT scans of the brain on the bone window, along with an axial brain CT scan on the brain window (D), reveal a well-defined bony defect in the left side of the clivus. The margins of the defect are well-defined, smooth, and devoid of any signs of aggression. A subtle, tiny, stalklike hyperdense structure projecting from the clivus is also visible, which represents a pathognomonic osseous stalk (arrows on B and D).

CT: computed tomography

MRI (Figure *2*) was performed with and without contrast. A well-defined midline cystic lesion measuring 6 × 10 mm was identified involving the dorsal aspect of the clivus. The lesion exhibited hypointensity on T1-weighted images, hyperintensity on T2-weighted images, and hypointensity following the cerebrospinal fluid (CSF) signal on fluid-attenuated inversion recovery (FLAIR) images. Diffusion-weighted imaging (DWI) revealed no evidence of restricted diffusion. Post-contrast T1-weighted images demonstrate no enhancement of the lesion. The remaining brain parenchyma appeared unremarkable.

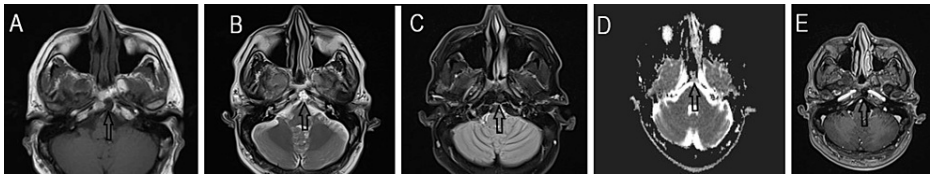


FIGURE 2: Brain MRI with and without contrast

The study demonstrated a well-defined midline clival lesion (arrow) measuring 6 × 10 mm cystic lesion with hypointense signal on T1-weighted images (A) and hyperintense signal on T2-weighted images (B) and hypointense signal on FLAIR image (C). In addition, the lesion did not show restricted diffusion (D) and did not demonstrate abnormal contrast enhancement on T1-weighted post-contrast images (E).

MRI: magnetic resonance imaging, FLAIR: fluid-attenuated inversion recovery

On reviewing the patient's medical file, an older MRI and CT imaging examination six years earlier was available; it showed a stable size and appearance of this small midline clival lesion. Based on the imaging features and characteristics, the diagnosis of EP was established. Notably, the lesion remained stable in size and appearance over a period of six years, reinforcing the benign nature of EP.

Discussion

Ecchordosis physaliphora (EP) is a benign lesion incidentally observed in skull base and spine imaging studies. It is a remnant of the notochord, a rod-shaped structure that is present during embryonic development and eventually forms the nucleus pulposus of the intervertebral discs [5]. EP is thought to occur in approximately 0.5%-2% of all autopsies and 1.5% of all brain MRIs but is often asymptomatic and goes unnoticed [1,6-8].

The diagnosis of EP is established based on characteristic imaging features of CT and MRI scans [3]. However, a CT scan is limited compared to an MRI in assessing EP for accurate localization and characteristics due to the small lesion size of EP and beam hardening artifacts in the posterior fossa images [1].

We presented a case of EP, which remains stable in size and appearance in imaging over a long period. The EP was incidentally discovered on a non-enhanced brain CT and enhanced brain MRI done in 2018, and the patient refused any further management and follow-up at that time.

On CT, EP typically appears as a well-demarcated, smoothly corticated bony defect in the clivus without aggressive features. Occasionally, a tiny, osseous, stalklike structure projecting from the clivus is considered

a hallmark and pathognomonic feature of EP on CT scans [3]. EP is usually hypointense on T1-weighted magnetic resonance imaging (MRI) and hyperintense on T2-weighted MRI. It does not typically enhance with gadolinium contrast. One distinguishing feature of EP is the presence of a thin, T2-hypointense pedicle that attaches the mass to the underlying clivus. This pedicle is thought to represent the remnant of the notochordal canal [1,2,5-9].

According to Ilorah et al., atypical features of EP include an absent bony stalk, a T2-hypointense protrusion from the clivus, T2 hypointensities bordering the lesion, a T2-hypointense center within the lesion, and a T2 hyperintensity on the pharyngeal surface or dorsum sellae [10].

The most important differential diagnosis for EP is intradural chordoma, a rare malignant tumor that also arises from notochordal remnants. Chordomas are typically larger and more aggressive than EP, with imaging features including bone destruction, mass effect, and contrast enhancement [1,4]. Other differential diagnoses for EP include lipoma, meningioma, arachnoid cyst, Tarlov cyst, epidermoid cyst, dermoid cyst, and metastatic tumor [11].

Furthermore, radiologists and physicians should be aware of the anatomic variants of the skull base, including the craniopharyngeal canal, arrested sphenoid pneumatization, and asymmetric pneumatization petrous apex, as all do not need intervention or further imaging follow-up [12-14].

EP is typically a benign incidental finding and does not require treatment. However, in rare cases, EP may cause symptoms such as headache, neck pain, or cranial nerve palsies. These symptoms are usually caused by mass effects on the surrounding structures. If EP is causing symptoms, surgical resection may be necessary [15].

Conclusions

The presented case of EP remained stable in size and appearance in imaging studies over an extended period. EP is a benign, incidental finding observed in imaging studies of the skull base and spine. It is crucial for radiologists and physicians to be familiar with the imaging characteristics of EP to accurately differentiate it from more aggressive lesions such as malignant chordoma. Follow-up examinations for EP patients would aid in establishing standardized diagnostic criteria and guidelines.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Yahea Alzahrani

Acquisition, analysis, or interpretation of data: Yahea Alzahrani

Drafting of the manuscript: Yahea Alzahrani

Critical review of the manuscript for important intellectual content: Yahea Alzahrani

Supervision: Yahea Alzahrani

Disclosures

Human subjects: Consent was obtained or waived by all participants in this study. The Research Ethics Committee (REC) of Armed Forces Hospital issued approval 2023-824. The Research Ethics Committee of Armed Forces Hospitals has reviewed the research project (2023-824) entitled "Case report of Ecchordosis physaliphora" and is pleased to inform you that this project has been approved under the expedited/normal procedure. The project complies with all requirements and policies established by the REC. Unless renewed, approval lapses one/two/three years after the approval date. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

References

1. Lakhani DA, Martin D: Ecchordosis physaliphora: case report and brief review of the literature . Radiol Case Rep. 2021, 16:3937-9. 10.1016/j.radcr.2021.09.049

2. Adamek D, Malec M, Grabska N, Krygowska-Wajs A, Gałązka K: Ecchordosis physaliphora - a case report and a review of notochord-derived lesions. Neurol Neurochir Pol. 2011, 45:169-73. 10.1016/s0028-3843(14)60029-3
3. Toda H, Kondo A, Iwasaki K: Neuroradiological characteristics of ecchordosis physaliphora. Case report and review of the literature. J Neurosurg. 1998, 89:830-4. 10.3171/jns.1998.89.5.0830
4. Mehnert F, Beschorner R, Küker W, Hahn U, Nägele T: Retroclival ecchordosis physaliphora: MR imaging and review of the literature. AJNR Am J Neuroradiol. 2004, 25:1851-5.
5. Ramesh T, Nagula SV, Tardieu GG, et al.: Update on the notochord including its embryology, molecular development, and pathology: a primer for the clinician. Cureus. 2017, 9:e1137. 10.7759/cureus.1137
6. Yamamoto T, Yano S, Hide T, Kuratsu J: A case of ecchordosis physaliphora presenting with an abducens nerve palsy: a rare symptomatic case managed with endoscopic endonasal transsphenoidal surgery. Surg Neurol Int. 2013, 4:13. 10.4103/2152-7806.106562
7. Macdonald RL, Cusimano MD, Deck JH, Gullane PJ, Dolan EJ: Cerebrospinal fluid fistula secondary to ecchordosis physaliphora. Neurosurgery. 1990, 26:515-8. 10.1097/00006123-199003000-00022
8. Stam FC, Kamphorst W: Ecchordosis physaliphora as a cause of fatal pontine hemorrhage. Eur Neurol. 1982, 21:90-3. 10.1159/000115460
9. Battal B, Zamora C: Imaging of skull base tumors. Tomography. 2023, 9:1196-235. 10.3390/tomography9040097
10. Ilorah C, Bond B, Kattah JC, Hassanzadeh B: An incidental finding of ecchordosis physaliphora in a case of abducens nerve palsy: case report. Neuroophthalmology. 2018, 42:233-6. 10.1080/01658107.2017.1381853
11. Nardi C, Maraghelli D, Pietragalla M, et al.: A practical overview of CT and MRI features of developmental, inflammatory, and neoplastic lesions of the sphenoid body and clivus. Neuroradiology. 2022, 64:1483-509. 10.1007/s00234-022-02986-x
12. Bello HR, Graves JA, Rohatgi S, et al.: Skull base-related lesions at routine head CT from the emergency department: pearls, pitfalls, and lessons learned. Radiographics. 2019, 39:1161-82. 10.1148/rg.2019180118
13. Thust SC, Yousry T: Imaging of skull base tumours. Rep Pract Oncol Radiother. 2016, 21:304-18. 10.1016/j.rpor.2015.12.008
14. Conley LM, Phillips CD: Imaging of the central skull base. Radiol Clin North Am. 2017, 55:53-67. 10.1016/j.rcl.2016.08.007
15. Sarkar N, Chakravarthy S, Chakravarty R, Mukhopadhyay S: Radiological diagnosis of a rare prepontine lesion: ecchordosis Physaliphora. Cureus. 2022, 14:e24335. 10.7759/cureus.24335